A simple method to ablate left-sided accessory pathways in a patient with coronary sinus ostial atresia and persistent left superior vena cava: A case report

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Introduction

Wolff-Parkinson-White (WPW) syndrome is characterized by a short PR interval, wide QRS complex, and paroxysmal supraventricular tachycardia.[1] An accessory atrioventricular connection is known to be the underlying mechanism of this syndrome. Accessory pathways (APs) are mainly located along the tricuspid or mitral annulus, accounting for 10%–20% and 50%–60% of cases, respectively.[2] Multielectrode recordings from a coronary sinus (CS) catheter facilitate the identification of the location of left free-wall APs; therefore, the usage of CS catheters is currently the standard for ablation of left free-wall APs.

Coronary sinus atresia is a rare anomaly and is generally accompanied by venous drainage into (1) a persistent left superior vena cava (PLSVC), (2) the left atrium through an unroofed CS, or (3) the cardiac chambers through collateral venous pathways.[3] This anomaly becomes a problem in procedures such as cardiac resynchronization therapy[4] or catheter ablation of a left-sided AP,[5] which require cannulation of the CS. Herein, we present a rare case of a 43-year-old woman with 2 left-sided APs accompanied by coronary sinus ostial atresia (CSOA) and a PLSVC.

Case report

A 43-year-old woman with intermittent WPW syndrome suffering from recurrent supraventricular tachycardia was referred to our hospital for a cardiac electrophysiology study (EPS) and ablation. The polarity of the delta waves was positive in all precordial leads with an R/S >1 in V_1, which suggested a left-sided AP. After written informed consent was obtained from the patient, an EPS was performed. Three multielectrode catheters were introduced from the right femoral vein and positioned in the high right atrium, His-recording region, and right ventricular apex. Then, coronary angiography was performed before the EPS, because the CS could not be cannulated from the right atrium. Coronary angiography showed ostial atresia of the CS and venous drainage into a PLSVC (Figure 1). Therefore, a left internal jugular vein puncture was performed and the sheath was inserted. The left-sided superior vena cava was directly cannulated via the left internal jugular vein, and a multipolar electrode catheter was positioned in the CS (Figure 2). The EPS suggested that an orthodromic atrioventricular reentrant tachycardia was the mechanism for the patient's tachycardia. Mapping of the mitral annulus was performed during ventricular pacing using a 7 French ablation catheter from the right femoral artery. The earliest retrograde atrial activation was recorded in the 4 o'clock to 5 o'clock region of the mitral annulus. Radiofrequency (RF) energy delivered to this site could not eliminate the bypass tract; however, the earliest retrograde atrial activation changed to the 2 o'clock to 3 o'clock region. The bypass tract was successfully eliminated by RF energy to this new site. Twenty minutes after the delivery of the RF energy, only ventriculoatrial conduction through the atrioventricular node remained. The 2 left-sided APs were smoothly ablated despite ostial atresia of the CS.

Discussion

We report a rare case of WPW syndrome coexistent with CSOA and a PLSVC. The incidence of CSOA is 0.03%–0.11%,[6,7] and more than half of cases with CSOA are accompanied by a PLSVC.[8] Although we described the venous drainage in the present case as a PLSVC, the venous drainage was smaller than a typical PLSVC with patent CS ostium. The small PLSVC size was attributable to the decreased flow volume. Coronary venous flow with CSOA was retrograded into the PLSVC. Therefore, the flow volume through the small PLSVC with CSOA was less than the flow volume through a typical PLSVC with the absence of a left innominate vein and CSOA. Decreased flow volume through a small PLSVC is associated with a small diameter, but is insufficient to obliterate it.[3]

KEYWORDS Wolff-Parkinson-White syndrome; Coronary sinus ostial atresia; Persistent left superior vena cava; Catheter ablation; Atrioventricular reentrant tachycardia (Heart Rhythm Case Reports 2016;0:1–4)

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KEY TEACHING POINTS

- Coronary sinus ostial atresia (CSOA) is a rare malformation and is accompanied by venous drainage into a persistent left superior vena cava (PLSVC) in more than 50% of cases.
- Coronary venous anomaly is more common in patients with atrioventricular reentrant tachycardia than in those with atrioventricular nodal reentrant tachycardia, possibly owing to an overlap in the stage (7–8 weeks of embryonic age) at which both the coronary sinus and accessory pathway develop.
- The direct cannulation of the PLSVC via the left jugular vein is a simple and effective strategy for managing this anomaly because accessory pathways in such patients are exclusively located in the left free wall.

CSOA matters during the ablation of a left free-wall AP because the multipolar electrode catheter located in the CS gives location information of the bypass tract between the left atrium and left ventricle (mitral annulus). However, the ablation of a left free-wall AP without a reference catheter has been reported from a few experienced centers. A previous report described a case with CSOA and a PLSVC, which was ablated by direct mapping of the mitral annulus with the ablation catheter.[5] Another previous report described a case of an AP positioned at the blind end of the CS (near the right atrium) coexistent with the same anomaly. A 6 French guiding catheter was introduced from the right femoral vein and into the CS via the left brachiocephalic vein and the PLSVC. The AP was successfully ablated from the right atrium under the guidance of a 2.5 French catheter with 16 electrodes through the guiding catheter in the CS.[9] Thus, the strategy mentioned above has been useful in a limited number of cases (ie, a case of an AP positioned at the blind end of the CS and a single left-sided AP). The left internal jugular vein was used for CS catheter access because the direct cannulation to the PLSVC via the left internal jugular vein was possible without the use of any devices, wires, or catheters. In previous reports,[4,9] the cannulation to the PLSVC via the left axillary or subclavian veins required the use of a Judkins right coronary catheter or a guiding catheter. We smoothly ablated 2 left-sided APs using the direct cannulation of a multipolar CS catheter from the left jugular vein across the left innominate vein into the PLSVC. Moreover, this method can be applied to any patient with a PLSVC, giving location information of APs, regardless of whether the CS ostium is atretic or not. The circulatory route in this case was as follows: venous blood from the CS went across the PLSVC into the left innominate vein and back towards the right atrium. Blood flow from the left jugular vein also converged into the left innominate vein, and rendezvous points of the venous drainage from the left jugular vein and PLSVC were near each other. Therefore, direct cannulation from the left jugular vein across the innominate vein to the PLSVC was possible.

A PLSVC is generally accompanied by undevelopment of the left innominate vein; therefore, venous blood from the left upper limb and head drains into the right atrium via the CS. The present case had a small PLSVC with retrograde flow, CSOA, and a left innominate vein. Coronary venous flow retrograded into the PLSVC. Blood flow of the left innominate vein was formed by the confluence of the left subclavian vein, the left jugular vein, and the small PLSVC. Venous blood from the left innominate vein went back toward the right atrium through the right superior vena cava. The points to be noted when mapping or ablating by this method are as follows: (1) a CS catheter does not inform the operator regarding the precise position of the mitral annulus; and (2) electrical potentials recorded from the PLSVC do not accurately reflect the potentials along with the mitral annulus. In a case without CSOA, a CS catheter is inserted from the right atrium to a great cardiac vein, which generally

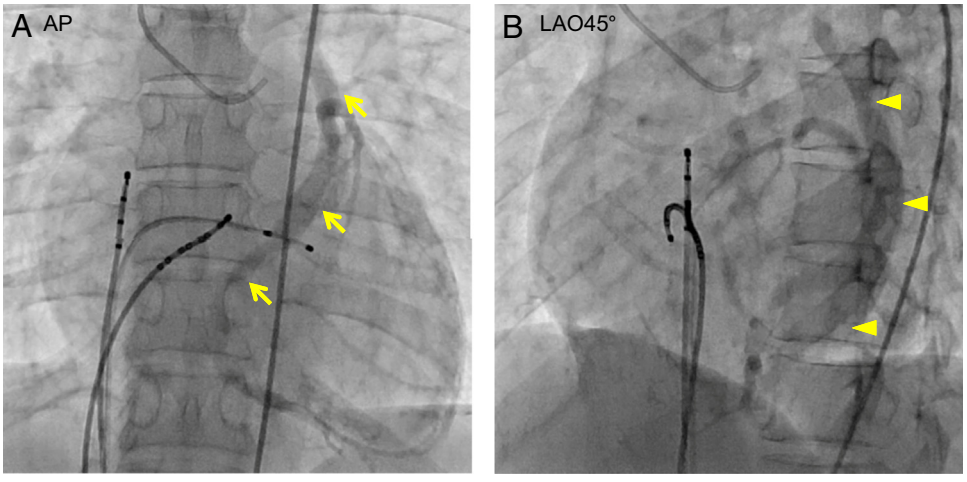

Figure 1 On the venous phase of the coronary angiography, there was no coronary sinus ostium and venous drainage into a persistent left-sided superior vena cava existed. **A:** Anterior-posterior (AP) view. **B:** Left anterior oblique (LAO) view. (A: *yellow arrows= PLSVC*) (B: *yellow arrowheads = PLSVC*)

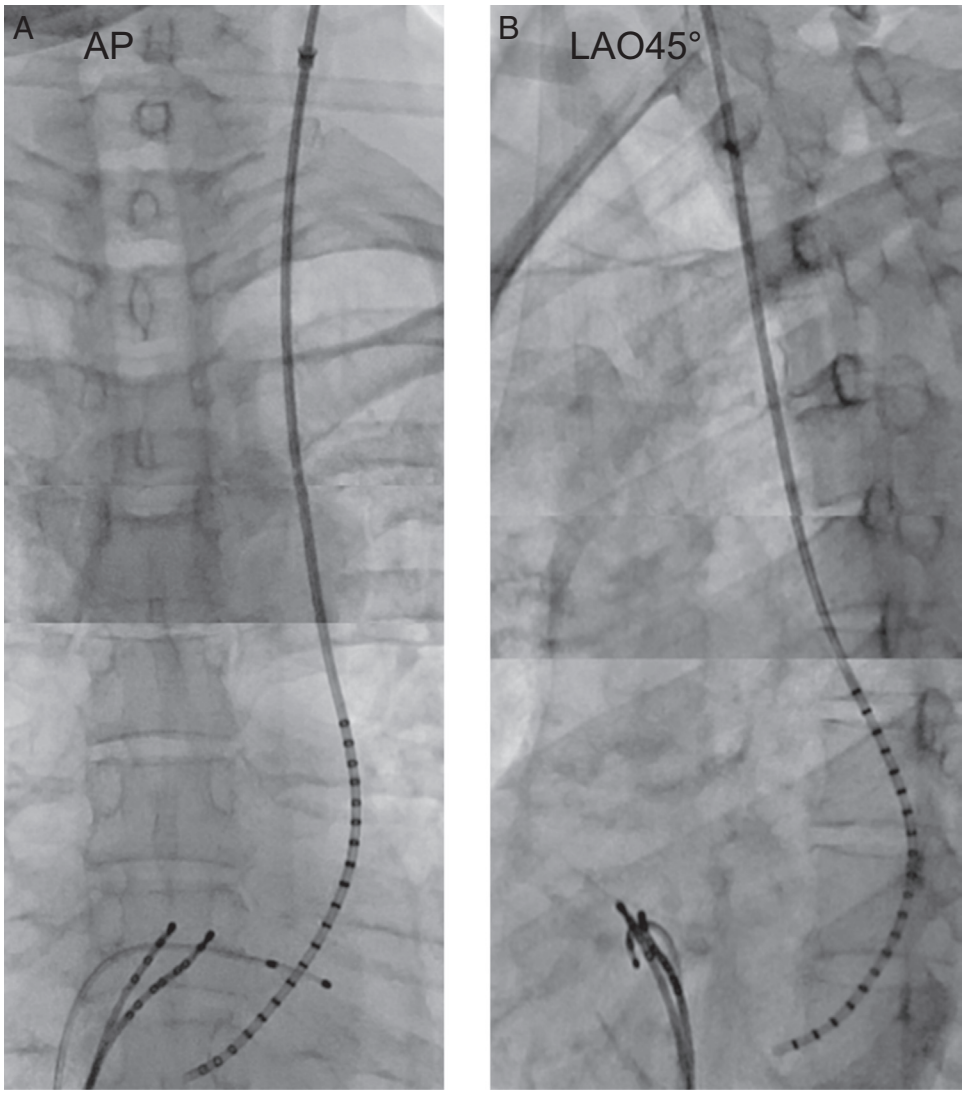

Figure 2 The direct cannulation of the multielectrode catheter from the left jugular vein to a persistent left-sided superior vena cava is shown. **A:** Anterior-posterior (AP) view. **B:** Left anterior oblique (LAO) view.

runs parallel to the mitral annulus. However, in the present case, the CS catheter was inserted via the PLSVC. The left cardinal vein differentiates into the ligament of Marshall, which courses from the CS obliquely above the left atrial appendage and lateral to the left superior pulmonary vein. Therefore, the PLSVC does not run parallel to the mitral annulus, because a PLSVC results from a failure of obliteration of the left common cardinal vein. In the present case, the CS catheter inserted via the PLSVC was located to the atrial side from the mitral annulus. Electrical potentials recorded by the CS catheter inserted via the PLSVC were different from electrical potentials of the mitral annulus, especially in the proximal side of the CS catheter. The proximal side of the CS catheter in the present case was the side opposite the atretic CS ostium near the right atrium. The distal side of the CS catheter was positioned in the blind end of the CS. Therefore, electrical potentials recorded in the distal side of the CS catheter almost matched the electrical potentials of the mitral annulus. On the other hand, electrical potentials recorded in the proximal side of the CS catheter did not reflect the electrical potentials of the mitral annulus. Although the earliest retrograde atrial activations were recorded in the CS3–4 region and the CS13–14 region, successful ablation sites were on the slightly proximal side of the CS catheter (ie, opposite the site of the blind end of the CS) from the earliest retrograde atrial activation recorded region. In addition, successful ablation sites included the slightly ventricular sides of the CS catheter positioned in the PLSVC (Figure 3).

An association between an AP and coronary venous anomaly was indicated in a previous report. APs result from incomplete fibrous separation of the atrial and ventricular myocardium. The CS develops from the proximal left sinus horn of the sinus venosus at 7–8 weeks of embryonic age, and an AP develops at the same stage. Chiang et al[10] reported that an anomaly of the CS was significantly more common in patients with atrioventricular reentrant tachycardia than in patients with atrioventricular nodal reentrant tachycardia. Moreover, APs in such patients are exclusively located in the left free wall and posteroseptal

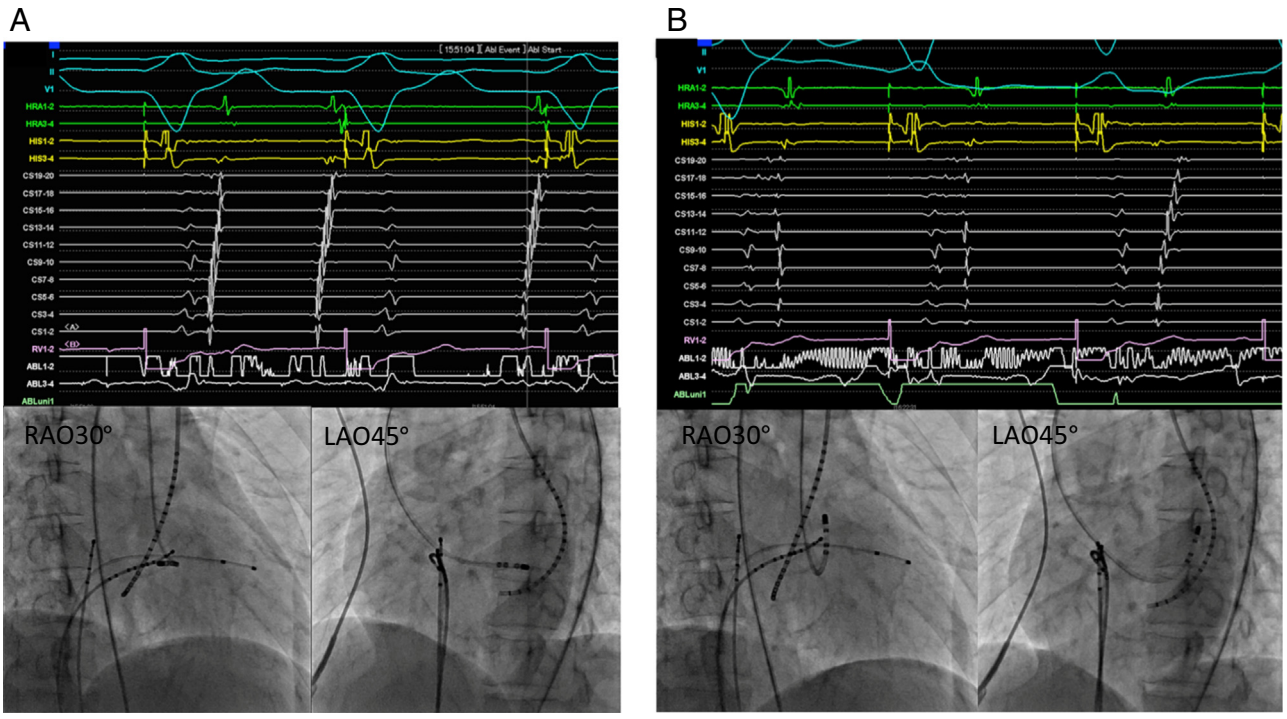

Figure 3 **A:** Although the earliest retrograde atrial activations were recorded in the coronary sinus (CS) 3–4 region, the successful ablation site of the first accessory pathway was on the slightly proximal side of the CS catheter (ie, opposite the site of the blind end of the CS) from the earliest retrograde atrial activation recorded region, and on the slightly ventricular sides of the CS catheter positioned in the persistent left superior vena cava (PLSVC). Using radiofrequency (RF) energy delivered to this site, ventriculoatrial (VA) conduction disappeared, but the retrograde atrial activation emerged again and changed to the CS13–14 region. **B:** Although the earliest retrograde atrial activations were recorded in the CS13–14 region, the successful ablation site of the second accessory pathway was on the slightly proximal side of the CS catheter (ie, opposite the site of the blind end of the CS) from the earliest retrograde atrial activation recorded region, and on the slightly ventricular sides of the CS catheter positioned in the PLSVC. After the delivery of the RF energy to this new site, only VA conduction through the atrioventricular node remained. LAO = left anterior oblique; RAO = right anterior oblique.

area. Prediagnosis of CSOA was another important issue in the current case. According to previous reports, angiography, computed tomography, and echocardiography are useful in detecting coronary venous anomalies. Diagnostic clues include a retrograde flow of a PLSVC, enlarged CS (>12 mm), and a small PLSVC (≤ 5 mm).[3]

In conclusion, the direct cannulation of the PLSVC via the left jugular vein made it possible to facilitate identification of 2 left free-wall APs in this rare malformation.

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